Preoperative diagnosis of bronchial atresia presenting with recurrent respiratory symptoms in an infant

Hasan Yüksel[1], Esen Demir[1], Remziye Tanaç[1], Recep Savaş[2], Hüdaver Alper[2]

Departments of [1]Pediatrics and [2]Radiology, Ege University Faculty of Medicine, İzmir, Turkey

SUMMARY: Yüksel H, Demir E, Tanaç R, Savaş R, Alper H. Preoperative diagnosis of bronchial atresia presenting with recurrent respiratory symptoms in an infant. Turk J Pediatr 2000; 42: 348-351.

Bronchial atresia (BA) is a rare respiratory malformation that may be diagnosed from infancy to adulthood. A typical feature of the disease is involvement of the left upper lobe and a mass-like lesion surrounded by a hyperlucent and nondeflating zone. We present a six-month-old male infant who was diagnosed by contrast-enhanced spiral computed tomography (CT) with three-dimensional (3D) technology, and by Tc-99m-macroalbumin aggregate (Tc99m-MAA) radionuclide scintigraphy. We stress that early diagnosis of BA can be made noninvasively using contrast-enhanced spiral CT and radionuclide scintigraphy. 3D computed tomographic reformation allows a more accurate diagnosis as well as a more specific approach to management and follow-up.

Key words: bronchial atresia, children, computed tomography, spiral.

Congenital bronchial atresia (BA) is a rare anomaly consisting of atresia or stenosis of a lobar or segmental bronchus, a mucocele formation behind the atretic area and a nondeflating hyperlucent segment of the lung[1]. The apicoposterior bronchus of the left upper lobe is almost always affected. However, right upper, right middle and, rarely, lower lobe involvement have been reported[2]. Although BA is a congenital malformation, it produces minimal symptoms and therefore may not become apparent until late childhood or even in adult hood[1-3].

So far, approximately fifty children have been reported in the literature. The incidence of BA has been reported as approximately two percent among children undergoing evaluation for bronchopulmonary malformation[4]. BA rarely causes recurrent respiratory problems. This anomaly is usually discovered on a routine chest roentgenogram[1]. We present an infant with BA presenting with recurrent respiratory symptoms, such as persistent cough, moderate respiratory distress, high fever and a persistent mass-like appearance at the left upper lobe of the lung.

Case Report

A six-month-old male infant was admitted to our clinic with persistent cough, moderate respiratory distress and persistent mass-like appearance at the left upper lobe of the lung on chest roentgenogram. His family had noticed these symptoms three months previously. Postero-anterior chest roentgenogram showed a mass-like lesion in the inspiratory phase, and an ipsilateral hyperlucent lung in the expiratory phase of breathing (Fig. 1). A finger-like mass produced by a mucus plug or mucocele at the apicoposterior segmental bronchus of the left upper lobe was detected with spiral computed tomography (CT) volumetric acquisition (using 3-mm collimation and a pitch of 1). This mucus plug was surrounded by multiple air-filled cysts and hyperlucent and oligemic lung tissue (Fig. 2). Three-dimensional (3D) surface rendering of the left upper lobe showed that it extended from the thoracic wall to the hilum at the apicoposterior segment of the left upper lobe (Fig. 3). Adjacent normal parenchyma was displaced away from the affected segment, and this pathological appearance extended down to the lower lobe of the left lung. Tc99m-macroalbumin aggregate (Tc99m-MAA) radionuclide lung perfusion scintigraphy revealed a total perfusion defect at the left apicoposterior segment of the lung (Fig. 4). The infant was treated with appropriate antibiotics

and supportive therapy. Left upper lobectomy was performed, and pathological examination of operation material revealed a segmental blind bronchus and a cyst-like intrabronchial plug, which was formed by inspissated mucus; the bronchial tree peripheral to the point of obliteration was patent and normal. Histological examination of the blind bronchus showed thinning of the bronchial wall and cartilage and a decreased number of alveoli, which were distended by inspissated mucus secretion.

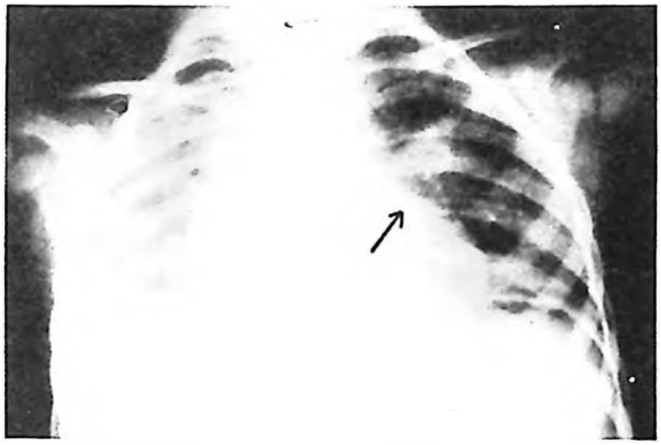

Fig. 1. Postero-anterior chest roentgenogram at expiration shows a mass-like lesion adjacent to the left hilum. The lung is hyperlucent and overinflated (note increased intercostal spaces on the left).

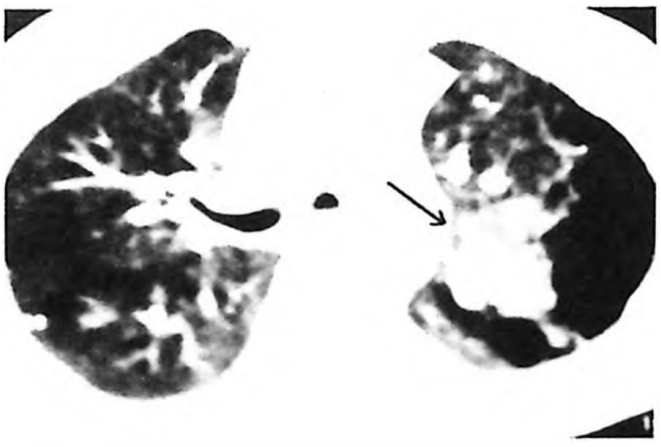

Fig. 2. Axial computed tomography (CT) scan shows mucoid impaction (arrow) with a hyperinflated apicoposterior segment of the left upper lobe.

Discussion

Bronchial atresia is a rare congenital malformation diagnosed in childhood[1,4]. The incidence of BA has been reported as approximately two percent among children under

Fig. 3. Three-dimensional (3D) surface rendering of the left upper lobe shows mucoid impaction clearly.

Fig. 4. Tc99m-macroalbumin aggregate (Tc99m-MAA) perfusion scintigraphy shows hypoattenuation in the left upper lobe (arrow).

going evaluation for bronchopulmonary malformation. Most of the reported patients are young children or adults because most affected infants or children have no significant clinical or recurrent symptoms showing respiratory problems[2,5,6]. Many patients having BA are diagnosed on routine chest roentgenogram without specific clinical symptoms. Thus, it may not become apparent until later childhood or even in adult hood. The involvement of the apicoposterior segment of the left upper lobe is the most characteristic feature[7]. The etiopathological origin is unknown. However, two theories have been postulated. The first is that the bronchial abnormality is probably developmental in origin. This theory was supported by some cases which were reported in early childhood, occasionally at birth, and with the concomitant presence of other congenital anomalies[8-10]. The second theory suggests that

a localized intrauterine interruption of the bronchial artery produces bronchial wall ischemia and secondary luminal obstruction[11].

Our patient had significant recurrent respiratory symptoms and moderate respiratory distress since birth. As mentioned above, rare cases are symptomatic in early childhood and thus can be diagnosed with a chest roentgenogram. A typical chest roentgenogram shows a mass-like appearance produced by a mucus plug, mucocele, and a hyperlucent, overexpended and nondeflating perimucocele zone. Adjacent normal parenchyma is compressed and depressed[1,2,12]. The inspiratory and expiratory roentgenograms reveal air-trapping behind the atretic segment. In our patient, routine images showed only a mass-like lesion (mucocele) in inspiratory images; expiratory roentgenogram showed nondeflating hyperlucent hemithorax typically. CT findings included multiple air-filled cysts, oligemia, a finger-like shadow demonstrating the mucus plug, mucocele, and hyperinflation in the affected segment[1,2,10,12].

Spiral CT is superior for showing mucus plug configuration, the finger-like shadow and the atretic segmental bronchus. Spiral CT images of our patient, obtained using reformatted images and enhanced 3D technology, showed that the finger-like shadow extended to the thoracic wall and was associated with a hyperinflated and hypoattenuated apicoposterior segment of the left upper lobe. The origin of the left apicoposterior segmental bronchus was not visible.

The mechanism of mucus plug formation is that retained fetal fluid and/or mucus secreted within patent airways distal to the point of atresia cannot pass the stenosis, and accumulate in the form of a plug[12]. The typical hyperlucent appearance points to the fact that the air can enter the affected segments only via collateral channels, creating an air draft between obstructed and adjacent normal ventilated segments and causing overinflation, expiratory air trapping and nondeflation[13]. Oligemia and hypoperfusion can be demonstrated using radionuclide perfusion scanning, as in our patient. We suggest that oligemia may be secondary to two causes. The first may be interruption of arterial supply as mentioned above. The other reason might be overinflation and displacement of the normal adjacent parenchyma, which may cause pressure on the pulmonary vessels and disruption in capillary circulation. This may affect the development of periatretic normal parenchyma and vasculature. Thus, early diagnosis and surgical treatment may help the normal development of the adjacent normal parenchyma. Our patient had a left upper lobectomy which improved his clinical condition. Respiratory symptoms did not recur for six months after surgery and he has been doing well since that time.

Bronchial atresia may be associated with bronchogenic cysts, anomalous pulmonary venous drainage and pectus excavatum[5,14]. Our patient had no other pulmonary or extrapulmonary malformations. Early surgical treatment should be considered especially in this situation.

Our experience showed that early diagnosis and approach to BA may be possible using both spiral CT and radionuclide perfusion scintigraphy before invasive procedures like bronchography and bronchoscopy. Spiral CT of the tracheobronchial tree with reformatted images, such as 3D technique, allows a more accurate diagnosis, and a more specific approach to surgical applications and follow-up of the cases. Therefore, evaluation of the tracheobronchial status of children with BA and associated respiratory anomalies might be possible using the above-mentioned technologies. Also, invasive procedures for demonstration of the tracheobronchial tree in children, especially in infants, may cause life-threatening complications and result in unsuccessful imaging. Thus, our proposed methods may lead to significant modification and selection of patient management, lower complications and also cost effectiveness.

In conclusion, BA should be considered in infants and children with a mass lesion, especially at the left upper lobe of the lung. Early diagnosis may be possible using enhanced spiral CT with secondary reformation and radionuclide perfusion scintigraphy. Spiral CT with secondary reformations may provide a more accurate approach to the medical and surgical management.

REFERENCES

1. Fraser R, Paré JA, Paré PD, Fraser R, Genereux GP. Pulmonary abnormalities of developmental origin. In: Fraser R, Paré JA, Paré PD, Fraser R, Genereux GP (eds). Diagnosis of Diseases of the Chest (3rd ed.) Vol. 2. Philadelphia: WB Saunders Co; 1989: 695-773.

2. Genereux GP. Bronchial atresia: a rare cause of unilateral lung hypertranslucency. J Can Assoc Radiol 1971; 22: 71-82.

3. Mori M, Kidogawa H, Moritaka T, Ueda N, Furuya K, Shigematsu S. Bronchial atresia: report of a case and review of the literature. Surg Today 1993; 23: 449-454.
4. Bailey PV, Tracy T Jr, Connors RH, deMello D, Lewis JE, Weber TR. Congenital bronchopulmonary malformations. Diagnostic and therapeutic considerations. J Thorac Cardiovasc Surg 1990; 99: 597-602.
5. Shishihara T, Tojima H, Fujita A, et al. [A case of congenital bronchial atresia complaining of chest pain with anomalous pulmonary venous drainage.] Nippon Kyobu Shikkan Gakkai Zasshi 1989; 27: 81-86.
6. Finck S, Milne EN. A case report of segmental bronchial atresia: radiologic evaluation including computed tomography and magnetic resonance imaging. J Thorac Imaging 1988; 3: 53-57.
7. Meng RL, Jensik RJ, Faber LP, Matthew GR, Kittle CF. Bronchial atresia. Ann Thorac Surg 1978; 25: 184-192.
8. Tsuji S, Heki S, Kobara Y, Sato A. The syndrome of bronchial mucocele and regional hyperinflation of the lung. Report of four cases. Chest 1973; 64: 444-447.
9. Haller JA Jr, Tepas JJ 3d, White JJ, Pickard LR, Robotham JL. The natural history of bronchial atresia. Serial observations of a case from birth to operative correction. J Thorac Cardiovasc Surg 1980; 79: 868-872.
10. Prinja N, Manoukian JJ. Neonatal/infant rigid bronchoscopy. J Otolaryngol 1998; 27: 31-36.
11. Robotham JL, Menkes HA, Chipps B, et al. A physiologic assessment of segmental bronchial atresia. Am Rev Respir Dis 1980; 121: 533-540.
12. Oh KS, Dorst JP, White JJ, Haller JA Jr, Jhonson BA,Byrne WD. The syndrome of bronchial atresia or stenosis with mucocele and focal hyperinflation of the lung. Johns Hopkins Med J 1976; 138: 48-53.
13. Culiner MM, Grimes OF. Localized emphysema in association with bronchial cysts or mucoceles. J Thorac Cardiovasc Surg 1961; 41: 306.
14. Khun C, Khun JP. Coexistence of bronchial atresia and bronchogenic cysts: diagnostic and embryologic considerations. Pediatr Radiol 1992; 22: 568-570.